



## International Journal of Surgery Case Reports

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## Ancient Schwannoma of superficial peroneal nerve presenting as intermittent leg pain: A case report



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## ARTICLE INFO

## Article history:

Received 8 August 2014

Accepted 8 November 2014

Available online 20 November 2014

## Keywords:

Ancient Schwannoma

Superficial peroneal nerve

Sheath tumors

Intermittent pain

## ABSTRACT

**INTRODUCTION:** Schwannomas are benign, encapsulated, slow-growing and usually solitary tumors originating from Schwann cells of the peripheral nerve sheath. Schwannomas of the superficial peroneal nerves are very rare, and therefore scarcely documented in the literature. The authors report a case of a diagnosed superficial fibular nerve sheath tumor with an unreported clinical presentation.

**PRESENTATION OF CASE:** A 52-year-old Caucasian female arrived to our Orthopedics Department complaining of pain and numbness of the lateral aspect of her left leg. These symptoms were present for a year and were more evident at the end of the day, or after a long time in the orthostatic position. No evidence of other medical illnesses was found. There was no record of prior traumatic events related to that limb. Diagnosis of a benign peripheral nerve tumor was achieved and the patient was treated by surgical excision of the lesion.

**DISCUSSION:** The intermittent symptomatology presentation on this case suggest a mechanical compression etiology, allied to classical pain and paresthesia often exhibited by this kind of the tumor. An intracompartmental pressure elevation could explain why the symptoms disclosed an episodic pattern, due to a constricted, inclosed nerve.

**CONCLUSION:** We describe a rare case of a patient with an unusual superficial peroneal nerve Schwannoma clinical presentation. Literature on this topic is scarce and, therefore, this case report intends to add further data about this kind of lesion.

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## 1. Introduction

Schwannomas, or neurilemmomas, are benign soft-tissue tumors, which arise from the Schwann cells of the peripheral nerve sheaths. Classically they are described as having a slow growth and uncommon malignant change. Although they represent the commonest benign peripheral nerve sheath tumors, the occurrence on the lower limbs account for 1% of all cases.<sup>1</sup> Reports of Schwannomas arising specifically from the superficial peroneal nerve are exceptionally rare.<sup>2</sup>

Ancient Schwannomas are rare variants of these tumors. They are termed “ancient” because of the degenerative features acquired with increasing age.

While usually clinically silent, they might be detected incidentally or become symptomatic due to mechanical compression and may present with pain, swelling or a lump. The diagnosis thus relies on the clinical history, physical examination, electrodiagnostic and imaging studies. Magnetic resonance (MR) neurography offers high-resolution visualization of structural peripheral nerve tumors, hence representing the diagnostic tool of choice when studying these kind of pathologic findings. The availability of MR neurography may be limited, and the costs can be significant, therefore ultrasonography may provide an economical and accurate imaging modality in diagnosis and management of these cases.<sup>3</sup>

Histologically, the characteristic degenerative findings of ancient Schwannomas may be mistakenly termed malignant. This type of benign mesenchymal neoplasia usually exhibits homogeneous nuclear hyperchromasia and pleomorphism with possibly small mitotic activity.<sup>4</sup>

Depending on the symptoms, surgical treatment is usually advised. Studies indicate that a neurilemmoma can be removed by

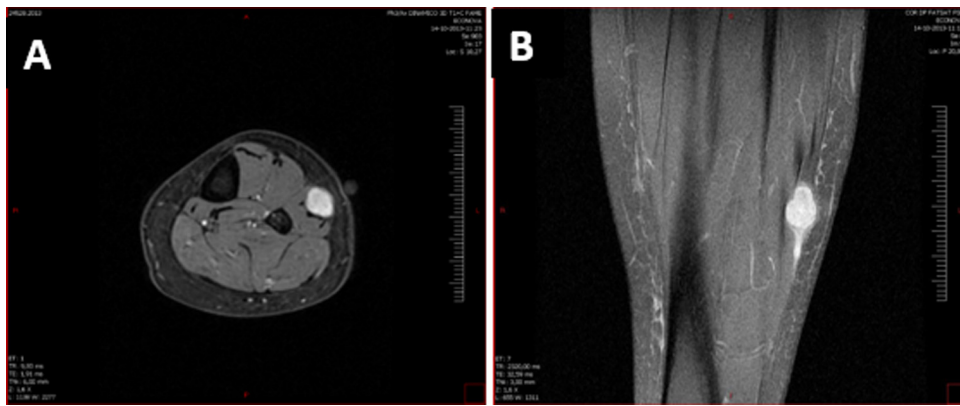
**Abbreviations:** MR, magnetic resonance; CT, computed tomography; EMG, electromyography; NCS, nerve conduction study.

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<http://dx.doi.org/10.1016/j.ijscr.2014.11.051>

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**Fig. 1.** Magnetic resonance imaging studies of the patient's left leg. Tumor visible on the lateral leg compartment below the level of the left fibular head. (A) Axial view on T1 weighted image. (B) Sagittal view on T1 weighted image showing a well-defined ovoid mass distal to the fibula head (measuring up to 18 mm in long axis), which arises, the underlying superficial peroneal nerve.

delicate enucleation with an acceptable risk of injury to the nerve trunk.<sup>5</sup>

Here, the authors present a case of a patient with an ancient Schwannoma located on the superficial peroneal nerve, presenting as intermittent leg and foot pain and paresthesia.

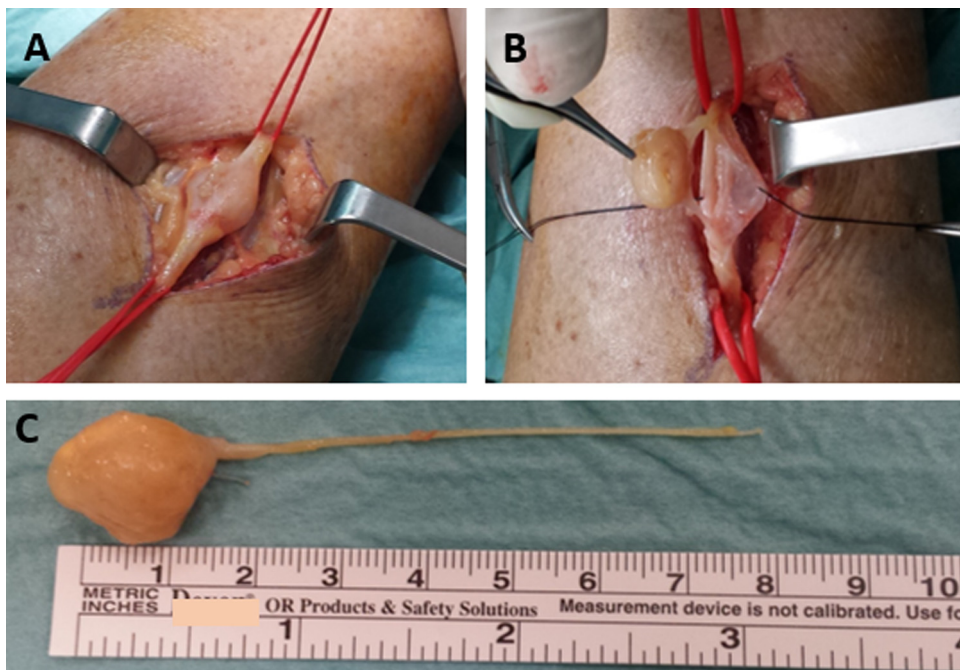
## 2. Case presentation

A 52-year-old Caucasian woman with no medical history arrived to the Orthopedic Emergency Department with a painful lump on her left leg. She denied any kind of traumatic event prior to the beginning of the symptoms. She referred a six months history of leg pain and numbness, and a gradually growing lump at her upper leg. The pain was moderate in intensity, burning in character and intermittent in nature. It irradiated distally and was made worse by crouching, crossing legs and long time standing. Clinical examination revealed a firm mass in the proximal lateral aspect of her left leg, measuring 3 cm × 2.8 cm, located on the subcutaneous plane

with little mobility in the coronal plane. It was not fluctuant and did not transilluminate. There was moderate tenderness in the same region. No other masses were found. She experienced increased pain with passive and active dorsiflexion. Motor function of the deep and superficial peroneal nerves was normal. However, with active foot inversion (with and without resistance), she experienced a non-well defined pain and sensory discomfort. By lightly tapping her leg lump, she revealed a positive Tinel's sign, eliciting a paresthesia on the dorsum of the foot.

Her investigations including blood results, D-dimer and venous ultrasound were unremarkable. Further evaluation was obtained by MR of the leg (Fig. 1). This imaging tool revealed a round to oval lesion in the peroneal compartment on the lateral aspect of left leg below the fibular neck, exhibiting intermediate signal intensity on T2 weighted images. These findings suggested a neof ormative lesion originating from the nerve sheath cells.

Surgical treatment was proposed, and the patient underwent intralesional excision by a peripheral nerve injury specialist.



**Fig. 2.** Surgical excision of the patient's left leg tumor. (A) Antero-lateral approach with tumoral exposure. (B) Perineural dissection and tumoral detachment of the nerve sheath. (C) Measurement of resected specimen.

The excised tumor (Fig. 2) was histologically studied and being described as a benign neurilemmoma with characteristics of ancient degenerative Schwannomas.

The postoperative period was uneventful, the patient's symptoms diminished considerably, and six months later she achieved complete healing and a symptom free state. She was able to walk without assistance and without pain or paresthesia.

### 3. Discussion

A Schwannoma, also known as neurilemmoma, neuroma, neurolemoma or Schwann cell tumor, is an encapsulated neoplastic lesion that arises from the neurilemmal cells which normally produce the insulating myelin sheath covering peripheral nerves.

Schwannomas most often occur in the fourth and fifth decade of life and seem to have a 1.6:1 female predilection.<sup>6</sup> They have been found to be present in such varied locations as the brachial plexus and the sciatic nerve. Rarely, Schwannomas can be found in the leg or in the foot and ankle region.<sup>7</sup>

Allied to the fact that they have a slow-growing pattern, they never traverse through the nerve but remain in the sheath lying on top of it. This explains why they are usually clinically silent and present oftentimes as an incidental finding. Patients may sometimes note cosmetic deformity or a palpable mass. Clear understanding of nerve structure and the peripheral nervous system aids diagnostic and therapeutic approaches to patients with peripheral nerve pathologic conditions.

In the present clinical case, it seems clear the symptomatology onset strongly correlates with the progressive tumor growth, whence voluminous enough to compress the adjacent nerve. This mechanical compression etiology is evident, since the pain and paresthesia were elicited or worsened in several conditions favoring this state, such as: the prolonged orthostatic position (facilitating edema of the lower extremity), crouching/crossing legs and tensioning and/or stretching the muscles of that anatomic region (active dorsiflexion, foot inversion). In all these situations an intracompartmental pressure elevation could explain why the symptoms disclosed an intermittent presentation, due to a constricted, inclosed nerve. It has been described that long periods of standing have been posited to be related to venous insufficiencies and leg fatigue,<sup>8</sup> discomfort and ultimately, pain.<sup>9,10</sup> Mechanical compressive forces cause cessation of retrograde and anterograde transport, further aggravating nerve dysfunction, structural damage and axonolysis, leading to Wallerian degeneration.<sup>11</sup>

The evaluation of a patient with a suspected pathologic nerve condition broadly includes history, physical examination, and ancillary studies including several imaging modalities which provide valuable information in these disorders. Plain films may not reveal any changes whereas computed tomography (CT), magnetic resonance (MR), especially MR neurography, may display a peripheral nerve tumor in a more detailed manner. Additional diagnostic tests, including electromyography (EMG) and nerve conduction study (NCS), evaluate neuromuscular function to assess denervation, preservation of motor units, or conduction loss.<sup>12</sup>

On microscopic analysis, Schwannomas appear solid or cystic and exhibit specific patterns: the Antoni A type has a spindle-cell array with collagen matrix arranged into palisading Verocay bodies; The Antoni B pattern exhibits a looser structure of mucinous matrix with fewer interspersed spindle cells; cellular Schwannomas display high cellular density and nuclear atypia, but contain fewer mitotic figures than malignant Schwannomas.<sup>12</sup>

The term "ancient Schwannoma" refers to the degenerative changes, which occur in these tumors with increasing duration. This rare variant was described by Ackerman and Taylor in 1951, and constitute 0.8% of all soft-tissue tumours.<sup>13</sup> The histologic analysis

is characterized by interstitial hyalinization, degenerative atypia, and cells with large and hyperchromatic nuclei.<sup>14</sup>

In this clinical case, radical surgical excision was advised due to the unendurable symptomatology. The intermittent pain was a day-to-day normal activity limiting condition.

Complete surgical resection of the tumor, whilst safeguarding the surrounding structures, resulted in cure, since no damage was sustained by the underlying nerve.

### 4. Conclusion

These benign neural sheath neoplasms are infrequent in the lower extremities according to the literature. In the differential diagnosis of nontraumatic leg pain, benign tumors, particularly Schwannomas of the peroneal nerves should be considered. Imaging studies, mainly magnetic resonance, are quite helpful in achieving a correct diagnose. Complete surgical resection by intralesional enucleation results in cure and recurrences are uncommon.

This case evinces a non-reported clinical presentation, on an infrequent location, for this specific neoplastic disorder, and so depicts an added value to the knowledge of these cases in the literature. Further studies are necessary to ascertain the etiology of the intermittent pain/paresthesia pattern presented by this patient.

### Conflict of interest

The authors declare that they have no competing interests.

### Funding

None.

### Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### Authors' contributions

All authors have been equally involved in the collection of data and in drafting the manuscript.

### Acknowledgements

The authors would like to thank Dr. Paulo Neves, Dra. Emilie Serrão and Dr. Henrique Salgado for their contribution to this case study.

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